

Well-trained sonographers are worth their weight in gold: ultrasound in systemic sclerosis

Some aspects of the role that ultrasonography (US) plays in rheumatic diseases are still a matter of debate. Although it is a non-invasive and real-time imaging modality, currently used the world over to assess disease activity as well as tissue and organ damage, it is still also frequently considered an operator-dependent technique in SSc. Indeed, doubts remain as to its utility, even if it provides a wide array of information for diagnosis and disease follow-up as well as for the quantification of lesions and damage at various sites. These sites include several structures and organs involved in SSc, such as the musculoskeletal system, lungs and skin, which are easily accessible for evaluation using US [1].

A high prevalence of synovial hypertrophy, joint effusion and bone damage was previously evidenced in SSc patients with arthralgia, with an 11–49% rate of power Doppler (PD) positivity at the wrists [2]. Interestingly, there was a similar prevalence of synovial hypertrophy and effusion when SSc patients were compared with a control group of RA patients, whilst bone erosions were more frequently observed in RA [3]. On the other hand, the presence of PD signals (particularly a mild grade) was reported to be similar in both populations [4]. Moreover, US demonstrated the presence of synovitis and tenosynovitis in a remarkable proportion of SSc patients (23 and 46%, respectively) with arthralgia without clinical signs of arthritis [5]. It has also been used to detect extra-articular abnormalities, particularly in patients with tendon friction rubs, where tendon and retinaculum thickening (mostly the A1 pulley) were more frequently observed in SSc individuals than in controls [3]. Furthermore, PD-positive tenosynovitis was a more frequent observation in the presence of tendon friction rubs, with tendon involvement that had a characteristic fibrotic pattern [3]. PD US in SSc patients may be also used to assess vasculopathy. A very recent study has also demonstrated that microvascular damage, evaluated by nailfold capillaroscopy, and macrovascular features such as ulnar artery occlusion assessed by PD are associated with the main digital manifestations of SSc, such as digital ulcers [6, 7].

Although there has been some interest in the use of US to assess the lung in SSc patients, there is a paucity of data in support of its advantages over high-resolution CT. Therefore, CT remains the gold standard diagnostic tool for interstitial lung involvement. Lung US is based on the detection of B lines, which are US artefacts generated by the thickened sub-pleural interlobular septa at the level of intercostal spaces, as well as on the evidence of sub-pleural nodules and pleural thickening. Although several studies have investigated the role of US in identifying lung

involvement in the past, only recently have validation studies been performed. It was demonstrated that semi-quantitative US scores have not only a good correlation with CT scores, but also a good reproducibility [8]. Interestingly, US can detect lung abnormalities before the onset of respiratory symptoms and prior to evidence of abnormalities in respiratory function tests [8]. These findings suggest that US might potentially become a support screening tool for the identification of SSc patients at risk of progressive lung involvement.

Progressive skin fibrosis is a characteristic of SSc and plays a central role in both disease subset classification and activity evaluation [1, 9]. The modified Rodnan skin score (mRSS) is able to distinguish patients with either limited (lcSSc) or diffuse cutaneous skin involvement [1, 9]. However, even if it is used as a primary outcome in most clinical trials, mRSS is unable to detect minimal skin thickness changes which, over time, might become clinically relevant [1, 9]. Recent studies have demonstrated that skin high frequency ultrasound can identify the oedematous phase that precedes palpable skin involvement in the first stages of SSc, facilitating early diagnosis [1, 10]. Indeed, high frequency ultrasound can detect subclinical diffuse dermal impairment in lcSSc patients that have a thicker dermis than healthy subjects in most skin areas with normal mRSS [1, 10]. These observations are also in agreement with recent microarray gene expression studies suggesting that clinically unaffected skin in SSc shares the peculiar gene signatures and pathology already evident in clinically affected skin [11]. A novel US application to analyse elastic properties of tissues is US elastography (UE). In patients with SSc, the first investigation by UE was made in 2010, after other studies demonstrated that UE can improve the reliability of US in measuring dermal thickness and ameliorate the assessment of fibrotic skin [12].

Another frequent finding in SSc patients is soft tissue calcifications, and in particular US is useful to study them at the level of the fingers, though today the best technique to assess these lesions remains radiography [13].

Even if it is able to access multiple sites in a single examination and both activity and damage can be evaluated, and despite the increasing evidence as to its value in SSc, US's diffusion as an assessment tool of this disease in clinical practice is still limited. A frequent matter of debate is the operator-dependence of US, which is usually reported as a major limitation of this tool in the assessment of various tissues and organs. However, when standardized scanning techniques and agreed definitions of pathology are used, US can be considered as reliable as any other imaging modality [14].

Then, what about US in SSc? On the basis of the current available data, US is a reliable imaging technique for the assessment of most musculoskeletal disorders (e.g. synovitis, tenosynovitis, bony erosions), if performed by an experienced ultrasonographer using standardized scanning techniques and agreed definitions of pathology. However, its reliability in other areas and organs, such as the skin and lungs, remains to be evaluated by further studies.

In conclusion, very promising data are currently present on the use of US for the assessment of lung and skin SSc involvement and more solid evidence is already available for the evaluation of the musculoskeletal system. We believe that well-trained sonographers are worth their weight in gold and provide further back-bone information to the physician in a complex disease such as SSc [2].

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